Airway Management and Central Venous Catheterization with Ultrasonography in a Patient with Wolf-Hirschhorn Syndrome

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Citation

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Abstract

One of the very rare chromosomal anomalies is the Wolf-Hirschhorn syndrome and can affect some parts of the body. The major features of this disease include a characteristic facial appearance, delayed growth and development, intellectual disability, and seizures. Syndromes with genetic disorders usually give difficult times to anesthesiologists. We report a 6-month-old boy with Wolf-Hirschhorn syndrome scheduled for US-guided central venous catheterization with laryngeal mask for airway management and vascular catheterizations in a pediatric patient with craniofacial abnormalities.

INTRODUCTION

One of the very rare chromosomal anomalies is the Wolf-Hirschhorn syndrome. These patients with this syndrome have deletion of the short arm of some chromosomes (chromosome 4). In these abnormality a characteristic facial appearance (distinctive facial features, including a broad, flat nasal bridge and a high forehead, widely spaced and protruding eyes, short philtrum, a downturned mouth, micrognathia,and poorly formed ears with small holes or flaps of skin, asymmetrical facial features and an unusually microcephaly), delayed growth and development, intellectual disability and seizures (1,2). And also have intrauterine growth restriction, severe psychomotor retardation, failure to thrive, profound developmental delays and various congenital midline fusion anomalies (3,4). We report a 6-month-old boy with Wolf-Hirschhorn syndrome scheduled for US-guided central venous catheterization with laryngeal mask for airway management and vascular catheterizations in a pediatric patient with craniofacial abnormalities.

CASE REPORT

A 6-month-old, 3 kg boy born with a prenatal diagnosis of Wolf-Hirschhorn syndrome was taken to the operation room for planned central venous catheterization. Wolf-Hirschhorn syndrome was diagnosed with the intrauterine ultrasound reports and following genetic testing. His physical exam was remarkable for micrognathia and eyes are widely spaced an done of protruding (Figure 1,2). Physical examination of the patient was noted to be small for his age, quiet and hypotonic. He had no prior surgical history. Preoperatively, respiratory rate: 25 breaths/min, oxygen saturation: 97% by pulse oximetry, heart rate: 140 beats/min, blood pressure (BP): 95/55 mmHg. Standard monitors were placed, and the patient underwent spontaneous breathing induction with oxygen, air and 5% sevoflurane. Mask ventilation was performed without difficulty and we inserted a size 1.5 laryngeal mask. After insertion of a laryngeal mask (see figure 3 below) anesthesia was maintained with sevoflurane. After the location of access was determined, local skin cleaning was performed with 10% povidone-iodine. The probe was covered with sterile glove and area of access was covered with sterile drape. The catheterization and puncture of the left internal jugular vein was performed through the ultrasound guided method, using a linear high frequency 6-18 MHz ultrasound probe (Esaote, MyLab™Five). During the intervention, the needle movements were followed on the US screen dynamically. When the blood flow into the injector in the vein became clear, the catheter was placed using the Seldinger method. Blood and fluid flow were checked using serum physiological administered through the catheter. Following the intervention, the patient was checked

for the position of the catheter and again for pneumothorax and hemothorax by chest radiography. The catheterization was performed uneventfully and the patient was transferred to the pediatric clinical unit with spontaneous breathing and in stable condition.

Figures 1-2

Images of the patient with Wolf-Hirschhorn syndrome with laryngeal mask and central venous catheterization.

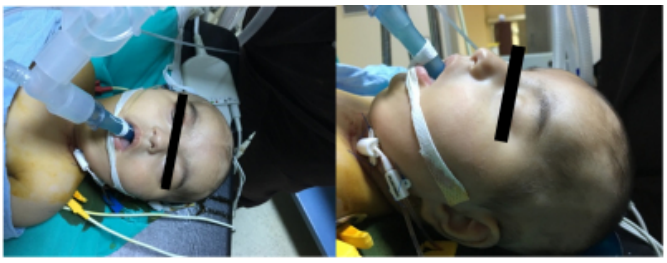
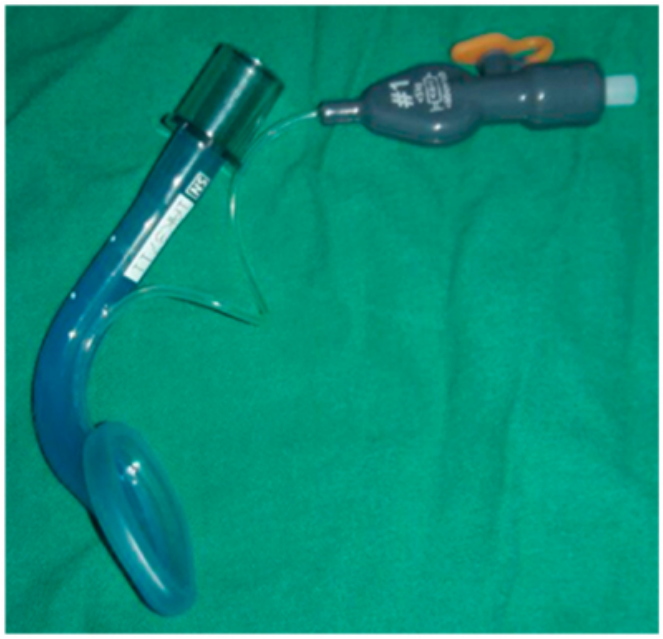


Figure 3

Image of the Laryngeal Mask



DISCUSSION

Children with genetic syndromes usually give difficult times to anesthesiologists (5). Babies with Wolf-Hirschhorn syndrome are born with congenital abnormalities that should be considered special when performing anesthesia, especially in the case of airway management. The potential perioperative complications of Wolf-Hirschhorn syndrome include facial anomalies (difficult airway management), congenital heart disease, hypotonia, seizure disorder, and the potential for chronic aspiration and perioperative respiratory problems. Few cases discussing the anesthetic management in patients with Wolf-Hirschhorn syndrome have been identified. In a case of a 33-month-old patient undergoing bilateral tympanoplasty and myringotomy, Choi et al. describe obtaining anterior, posterior, and lateral radiography of the cervical spine, in addition to performing a standard airway evaluation to assist in the identification of airway abnormalities (6). Two cases describe the use of downsized endotracheal tubes, either as a result of difficulty with intubation or in an attempt to minimize the potential for airway challenges (6,7). None of the cases reviewed describes central venous catheterization with ultrasonography in a patient with Wolf-Hirschhorn Syndrome. We describe a successful laryngeal mask insertion and central venous catheterization using ultrasonography in such a patient.

CONCLUSION

Wolf-Hirschhorn Syndrome has an important place in the management of anesthesia in terms of airway difficulty. For these patients, all preoperative airway materials should be available. It is also difficult to insert central venous catheters because of the short neck. Ultrasonographic help is warranted and no such case was found in the literature.

References

1. Hirschhorn K, Cooper HL, Firschein IL. Deletion of short arms of chromosome 4-5 in a child with defects of midline fusion. Humangenetik. 1965;1(5):479-482.
2. Wolf U, Reinwein H, Porsch R, Schroter R, Baitsch H. [Deficiency on the short arms of a chromosome No. 4]. Humangenetik. 1965;1(5):397-413.
3. John F. Gamble, Dinesh J. Kurian, Andrea G. Udani, and Nathaniel H. Greene. Airway Management in a Patient with Wolf-Hirschhorn Syndrome. Hindawi Publishing Corporation Case Reports in Pediatrics Volume 2016, Article ID 7070125, 2 pages http://dx.doi.org/10.1155/2016/7070125.
4. Humston C, Bernard R, Khan S, Tobias J.D. Perioperative Care of an Infant With Wolf-Hirschhorn Syndrome: Is There a Risk of Malignant Hyperthermia. Journal of Medical Cases. Volume 7, Number 4, April 2016, pages 126-129.
5. Butler MG, Hayes BG, Hathaway MM, Begleiter ML. Specific genetic diseases at risk for sedation/anesthesia complications. Anesth Analg. 2000;91(4):837-855.
6. J. H. Choi, J. H. Kim, Y. C. Park, W. Y. Kim, and Y.-S. Lee, "Anesthetic experience using total intra-venous anesthesia for a patient with Wolf-Hirschhorn syndrome—a case report,"Korean Journal of Anesthesiology, vol. 60, no. 2, pp. 119–123, 2011.
7. R. Ginsburg and G. Purcell-Jones, "Malignant hyperthermia intheWolf-Hirschhorn syndrome," Anaesthesia, vol. 43, no. 5, pp. 386–388, 1988.

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